Sturge-Weber-Dimitri Disease: Role of Hemispherectomy in Prognosis

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ABSTRACT: We report 12 patients with Sturge-Weber-Dimitri disease treated surgically between January 1975 and December 1987. Hemispherectomy was performed on ten, two others underwent occipital lobectomy for intractable seizures. All operations were performed between the ages of 3 months and 20 months, except in two at age 8 and 9 years. The onset of seizures in all was between 2 and 8 months of age, except for two at 15 months. There were no postoperative deaths. Postoperative shunt procedures were required in 3 out of 12 (25%). Postoperative seizure control for one year or more was achieved in 11 out of 12 patients. The remaining patient is on medication with no seizures, but follow-up is less than a year. Intellectual deterioration was not seen after surgery except in 2 patients who had late operations. We conclude that patients with Sturge-Weber-Dimitri syndrome who have intractable seizures in the first 6 months of life and unilateral hemispheric involvement should be considered for early resection of the involved hemisphere.

RÉSUMÉ: Le rôle de l'hémisphérectomie dans le prognostic de la maladie de Sturge-Weber-Dimitri Nous rapportons les cas de 12 patients atteints de la maladie de Sturge-Weber-Dimitri qui ont été traités chirurgicalement entre janvier 1975 et décembre 1987. Une hémisphérectomie a été effectuée chez 10 patients et 2 autres ont subi une lobectomie occipitale pour épilepsie réfractaire au traitement. Toutes les interventions ont été réalisées entre l'âge de 3 et 20 mois, sauf chez deux patients âgés de 8 et 9 ans respectivement. Le début des crises convulsives se situait entre 2 et 8 mois chez tous les patients sauf chez deux qui avaient commencé à 15 mois. Il n'y a eu aucun décès en post-opératoire. On a dû avoir recours à un shunt post-opératoire chez 3 des 12 patients (25%). On a obtenu un contrôle des crises pendant un an ou plus après l'intervention chez 11 des 12 patients. L'autre patient n'a pas eu de crise sous médication après un suivi de moins de 12 mois. Nous n'avons pas observé de détérioration intellectuelle après la chirurgie sauf chez 2 patients opérés tardivement. Nous concluons que, chez les patients atteints du syndrome de Sturge-Weber-Dimitri qui présentent des crises convulsives réfractaires au traitement dans les premiers six mois après la naissance et dont l'atteint hémisphérique est unilatérale, une résection précoce de l'hémisphère atteint doit être considérée.

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Sturge-Weber-Dimitri disease (SWD) is an uncommon condition first described by Sturge in 1879 in a case report of an epileptic patient.[1] The association of intracranial calcifications was subsequently described by Weber in 1922[2] and Dimitri in 1923[3], giving this condition its familiar eponym.

The classical findings include gyriform intracranial calcifications, congenital and usually unilateral port-wine naevus of the skin of the face in the territory of one or more divisions of the trigeminal nerve on the same side as the cerebral lesion, convulsive disorder, glaucoma and some degree of deterioration of mentation and hemiparesis.

Variants of the classical findings have been described in the literature such as absence of facial port-wine stain,[4,5] extensive intracranial calcifications contralateral to the bulk of the facial naevus,[6] severe bilateral skin involvement with only unilateral cerebral involvement,[7] facial port-wine stains and glaucoma without meningeal anomalies,[8] ocular anomalies involving the scleral vessels, choroid, or retina[9] and associated hemiatrophy or hypertrophy of limbs.[10]

SWD can be clinically confirmed with contrast-enhanced computed tomographic scanning (CT) which shows the distribution of the calcification in a gyral pattern, hemicerebral atrophy and angiomatous enlargement of the choroid plexus on the involved side as described by Stimac et al in 1986.[11]

If seizures begin in the first months of life, there appears to be a high risk of progressive mental deterioration. Alexander and Norman,[14] in their studies on this disease showed that if seizures began in infancy, then by later childhood the patient would be institutionalized because of dementia and hemiplegia and would most probably die in status epilepticus at an early age. They recommended that surgical intervention in this disorder be carried out early in infancy. The purpose of this study was to examine the clinical profile, prognostic factors, and role of hemispherectomy (decorticectomy) in SWD.

Materials and Methods

We reviewed the hospital records of 12 patients with SWD treated surgically between January 1975 and December 1987 at The Hospital for Sick Children, Toronto, and recorded specific information such as age, sex, naevus distribution, involved hemisphere, age at onset of seizures, response to antiepileptic drugs, presence of glaucoma, CT findings, age at decortication,

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postoperative shunt requirements, Intelligence Quotient (IQ) or developmental assessment before and after surgery, electroencephalogram (EEG) preoperatively, postoperative seizure control, and duration of follow-up.

The postoperative seizure control at one year or more on follow-up was denoted by six grades, shown in Table 1 as adapted from J. Kobayashi et al.[12]

Table 1: Post Operative Grades of Seizure Control

Grade I	Seizure-free without medication
Grade II	Seizure-free on anticonvulsant drugs
Grade III	> 50% reduction in seizure frequency
Grade IV	< 50% reduction in seizure frequency
Grade V	Not improved
Grade VI	Worse

RESULTS

Twelve patients had SWD during the study period, with a female preponderance of 10 and 2 males; ages ranged at operation between 3 months and 9 years (Table 2). All operations were performed between 3 months and 20 months of age, except two patients who had surgery at 8 and 9 years. Ten patients underwent hemispherectomy and 2 had localized excision of the occipital lobe. All the operated patients had intractable seizures refractory to anti epileptic drugs. The seizure type in all patients was partial motor, except in one patient who initially had elementary partial motor seizures later evolving into complex partial seizures. The onset of seizures in all patients was between 2 and 8 months, except in two whose onset was at 15 months. Ten of the 12 patients had progressive hemiparesis, including all those with diffuse hemispheric involvement on CT scan. Two patients with only partieto-occipital involvement did not have hemiparesis. The EEG of all

Table 2: Patients with Sturge-Weber-Dimitri Syndrome Who Have Undergone Cortical Excision at The Hospital for Sick Children

Patient No.	Naevus Distribution	Glaucoma	Age at Onset of Seizures	Clinical Type of Seizure	CT Cerebral Involvement	Age at Operation	Type of Operation (S-Shunt)	Post-Operative I.Q.	Post-Operative Seizure Control (Grade)	Post-Operative Follow-up (years)
1	LV_1V_2	None	6 months	R partial motor	L hemispheric	10 months	L hemispherectomy (S)	92	I	12
2	RV_1V_2 LV_1V_2	Yes R > L	4 1/2 months	"	" plus R frontal	5 months	" (S)	93	II	10
3	$RV_1V_2V_3$ $LV_1V_2V_3$	Yes L	2 1/2 months	"	bilateral hemispheric L > R	9 years	"	52	II	2
4	$LV_1V_2V_3$	Yes L	4 months	"	L hemispheric	6 months	" (S)	99	II	11
5	RV_1V_2	Yes R	2 months	L partial motor	R hemispheric	3 months	R hemispherectomy	93	I	11
6	LV_1V_2	Yes L	15 months	R partial motor	L occipital lobe	20 months	L occipital lobectomy	88	I	5
7	$RV_1V_2V_3$ $LV_1V_2V_3$	Yes R	4 1/2 months	L partial motor	bilateral R > L	5 months	R hemispherectomy	93	I	4
8	$RV_1V_2V_3$ $LV_1V_2V_3$	Yes R > L	8 months	R partial motor	bilateral L > R	11 months	L hemispherectomy	90	I	3
9	RV_1V_2	None	2 1/2 months	L partial motor	R hemispheric	3 months	R hemispherectomy	99	I	12
10	RV_1V_2	None	3 months	"	"	12 months	"	92	II	10
11	$LV_1V_2V_3$	Yes L	15 months	R partial motor	L occipital lobe	20 months	L occipital lobectomy	90	II	9
12	$LV_1V_2V_3$ RV_2V_3	"	8 weeks	Initially R partial motor → CPS	L hemispheric	8 years	L hemispherectomy	40	II	1

R - Right
L - Left
CPS - Complex partial seizure

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patients showed changes referable to the involved hemisphere, such as attenuation of background activity with or without epileptiform abnormalities. SPECT scanning with 99_{Tc} Hm-PAO showed marked increase in regional cerebral blood flow coextensive with the intracranial involvement in the 4 patients in whom this investigation was performed.[13] Computed tomography showed either localized or diffuse hemispheric calcification, enlarged choroid plexus with moderate to severe cerebral atrophy on the involved side in all patients.

Nine out of 12 of the operated patients (75%) had associated glaucoma ipsilateral to the skin lesions requiring therapy.

There were no postoperative deaths in any of the decorticated patients. Postoperative follow-up ranged between 1 and 12 years, with a mean follow-up period of 8.3 years. Postoperative shunt procedures were performed in 3 of 12 patients (25%); two developed communicating hydrocephalus requiring ventriculoperitoneal shunt, and one developed a subdural fluid collection and required a subdural-peritoneal shunt. All became shunt-dependent. Postoperative seizure control at one year or more was achieved in 11 of 12 patients: six were on no medication (Grade I), five were on medication with no seizures (Grade II). One patient is on medication and free of seizures, but follow-up is less than one year. Improvement of intelligence of varying IQ (82-99) was seen postoperatively on follow-up in all except in two, who had operations late. In 8 patients who had early hemispherectomy (less than 12 months) the mean IQ was 94. In 2 patients with late hemispherectomy (more than 12 months) the mean IQ was 46. In 2 patients who had occipital lobectomy, the mean IQ was 89 (Table 2). The hemiparesis in patients who were operated early resolved almost completely, although 2 patients who had late operations showed no improvement in motor function.

Discussion

The clinical course of SWD varies, but Alexander and Norman showed that if seizures begin in early infancy, there appears to be a high risk of progressive mental deterioration with a downhill course of severe hemiplegia and intractable seizures.[14]

The venous angioma of the SWD leads to reduction of blood flow through the involved hemisphere with consequent cerebral atrophy and seizures affecting the contralateral uninvolved hemisphere, impairing its function and leading to severe mental retardation. Prompt surgical treatment as originally advocated by Alexander and Norman[14] allows for preservation of function of the normal remaining hemisphere.

Lesions restricted to the occipital lobe tend to produce fewer seizures and delayed seizure onset,[15] as seen in 2 of the patients (Cases 6 and 11) who exhibited an onset of seizures at 15 months, compared to the rest with onset between 2 and 8 months.

Our postoperative results are encouraging. All operations performed between the ages of 3 and 20 months had good intellectual development, compared with operations performed at age 8 and 9 years, who were already severely retarded. However, cessation of seizures was achieved in all patients with or without anticonvulsant drugs. The degree of hemiparesis in SWD is less than in children who incur hemispheric insults beyond infancy. Ramirez de Arellano[16] in 1961, found that monkeys subjected to hemidecortication at age 6 days had excellent preservation of function in the contralateral limbs and face as compared to monkeys who had their decortication at age 19 months. Hoffman et al[17] in an earlier report found that early decortication helps preservation of function of the normal remaining hemisphere, cessation of seizures, less severe of hemiparesis, and maintenance of reasonably good intelligence.

In conclusion, patients with SWD who have partial motor seizures in the first 6 months of life, progressive hemiparesis and diffuse hemispheric involvement on CT Scan should be considered for early hemispherectomy.

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